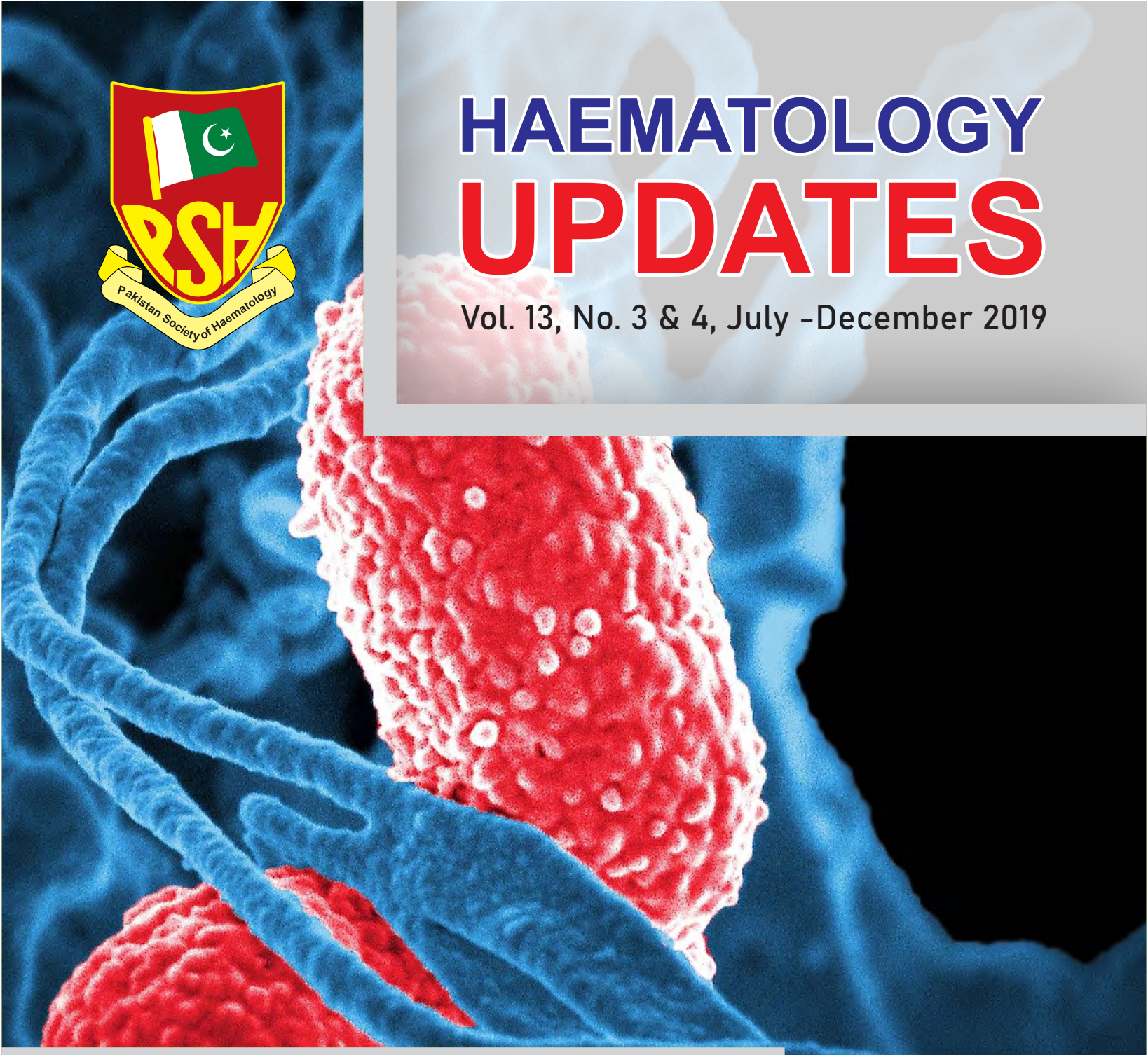
# HAEMATOLOGY UPDATES

Vol. 13, No. 3 & 4, July -December 2019



## IN THIS VOLUME

# PRESIDENT'S COLUMN

Pakistan Society of Haematology (PSH) has been very active during the previous and ongoing quarters, has organized lot of academic seminars and workshops that has made tremendous contributions not only by enhancing haematology human resource development but also creating awareness among the medical fraternity all over the country.

Professor Nisar Ahmed and his team at Children's Hospital and Institute of Child Health, Lahore is organizing PSH mega event 22nd Annual Meeting (HaemCon 2020) from 13-16 February 2020 at Avari Hotel Lahore. A large number of national and international experts will be giving talks and state of the art lectures. I will indeed provide an invaluable educational experience and opportunity to interact and share experience with experts from across the world. The highlights of this meeting are exclusive session on Haem-Onc Nursing and young researcher's awards.

We look forward to welcome you all in the HaemCon2020.

Yours sincerely,

**Dr Parvez Ahmad**
President, Pakistan Society of Haematology (PSH)
Professor of Clinical Haematology & HOD
Quaid-e-Azam International Hospital, Islamabad - Pakistan

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# JOIN US!

Assalam-o-Alikum Dear Colleagues,

The month of December 2019 was very gloomy for me as well as for Pakistan Society of Haematology (PSH) because our beloved fathers left for their heavenly abode. إِنَّا لِلّٰهِ وَإِنَّـا إِلَيْهِ رَاجِعونَ

My own father Mr. Iqbal Ahmed Ch and father of Haematology in Pakistan Lt Gen Muhammad Saleem, HI(M) will always be remembered for their legacy & selfless services.

May ALLAH Almighty bless them with higher ranks in heavens and give us the courage & honours to follow their footsteps.

مقدور ہو تو خاک سے پوچھوں کہ اے لئیم
تو نے وہ گنج ہائے گراں مایہ کیا کیے

**Fathers Never Die,** They just go on a long leave. Their persona lives on and their words keep on echoing. Their experienced advices & guidance gathered over years of hard work are easiest to find and their footsteps are always available to be followed. Perhaps the word FATHER is synonym for immortality!

*Your life was a blessing*
*Your memory a treasure*
*You are loved beyond words*
*And missed beyond measure*

*We haven't really left you*
*Even through it may seem so*
*We have gone to our heavenly home*
*And am closer than you know!*

Mr. Iqbal Ahmed Ch. (1937 - 2019)

Lt Gen (R) M Saleem, HI(M) (1937 - 2019)

Wishing sincerely for the health, happiness & prosperity of all members and their lovely families. See you all in HAEMCON2020 Lahore.

Your sincerely,

**Dr Mehreen Ali Khan**
Secretary, Pakistan Society of Haematology (PSH)
Consultant Haematologist & BMT Specialist
AFBMTC/NIBMT, Rawalpindi - Pakistan

# ABOUT PSH

Pakistan Society of Haematology (PSH) was formed in 1996 with the aim of promoting advancement of haematology, BMT and transfusion medicine in the country. Presently it has more than 500 members and we all should make efforts to enroll every haematologist in the country. We request all our members to take special interest in extending the membership to all those haematologists around you who have not yet registered with PSH. Website was launched and has been very active in recent past. We are trying to rejuvenate the website "https://www.psh.org.pk. The website would be interactive and provide on line forum for sharing views with other haematologists, and case discussion with the experts. Other features will be facility to download online membership form, newsletter, list and addresses of the members. Hopefully the website will be more operational within this month InshaALLAH.

**History:**
PSH was raised as "Pakistan Society of Haematology/Transfusion Medicine (PASHT)" in 1991. A meeting was held at 5 pm on Friday Nov 22, 1991. Professor Dr Mohammad Khurshid, Brig(later Lt Gen) Muhammad Saleem, Dr Khalid Zafar Hashmi, Dr Nasim Siddiqui, and Dr Abdul Hayee attended the meeting as members in presence of Prof A. V Hoffbrand. In this meeting Dr Khurshid presented a brief outlay of the necessity to create such a society. He also pointed out that Dr. Abdul Hayee, Dr. Khurshid, Dr KZ Hashmi and Brig Saleem had met at Bahawalpur and agreed on the general principles that the first meeting would be held along with the International conference of Pathology

Though initial work was comprehensive, governing body and meetings of PASHT were not held regularly. In Sept 1994 it was proposed by Gen Muhammad Saleem to meet all PASHT members during Pakistan Association of Pathology (PAP) conference at Quetta. Dr. Muhammad Khurshid in consultation with Gen Saleem, Prof. Abdul Hayee, Dr. Khalid Zafar Hashmi proposed a provisional constitution of PASHT for the discussion in meeting.

Haematologists from all over the country met on Saturday 9 March 1996 at Hotel Pearl Continental Rawalpindi in order to form a society. It was unanimously agreed that official name of society will be "Pakistan Society of Haematology" with official abbreviation of "PSH". It was also decided that until elections for office bearers the society matters will be looked after by a committee as under

a. Dr. Muhammad Khurshid
b. Dr. Ehsan-ul-Allah
c. Dr. Abdul Hayee
d. Dr. Khalid Zafar Hashmi
e. Dr. Khalid Hassan
f. Dr. Masood Anwar will act as Co-ordinator

A general body meeting of PSH was held at Peshawar on 2 and 3 Nov 1996. Election for office bearers were carried out as follow

# ABOUT PSH

a. Lt. Gen. Muhammad Saleem President
b. Prof. Muhammad Khurshid as Vice President
c. Dr. Khalid Hassan as Secretary/treasurer

Later in Oct 1997 appointment of vice president was renamed as president elect.

List of past presidents includes

1. Prof. Dr. Abdul Hayee
2. Prof. Dr. Abdul Khaliq
3. Prof. Dr. Muhammad Khurshid
4. Prof. Dr. Khalid Zafar Hashmi
5. Maj. Gen. Masood Anwer
6. Prof. Dr. Khalid Hassan
7. Maj. Gen. Suhaib Ahmed
8. Prof. Dr. Samina Naeem
9. Maj. Gen. Muhammad Ayyub
10. Prof. Dr. Nisar Ahmed

List of past secretaries includes

1. Dr. Khalid Hassan
2. Col. Massod Anwar
3. Prof. Fazle-e-Raziq
4. Dr. Salman Naseem Adil
5. Dr. Shaheena Kauser
6. Brig. Nadir Ali
7. Brig Pervez Ahmed
8. Dr. Nadeem Ikram
9. Dr. Humera Rafiq
10. Brig. Tariq Mehmood Satti
11. Dr. Saima Farhan

PSH was registered with Govt of Pakistan on 8 August 1998(RS/ICT/298 dated 8 Aug 1998 as non political and non sectarian body to promote advancement of haematology including transfusion medicine through encouragement of research, teaching and technical methods. The body will also organize scientific meetings, publication of scientific material, and affiliation with other National and international organizations. Members of Governing body included

a. Lt. Gen. Muhammad Saleem as President
b. Dr. Khalid Hassan as General secretary
c. Dr. Birgees Mazhar Qazi as member
d. Dr. Waseem Iqbal as member
e. Dr. Hassan Abbas Zaheer as member
f. Dr. Mobina Ahsan Dhodhy as member
g. Dr. Farah Yasin as member
h. Col. Masood Anwar as member

It was also decided that first National conference will be held on 4 Oct 1998. Since then Annual conference is held regularly in all capital cities of Pakistan. The society is publishing a quarterly newsletter and providing a forum to the haematologists all over the country contributing as advisors in haematology, consultants, researchers and mentorship. Currently the Governing body includes

Prof. Dr. Parvez Ahmed as President
Prof. Dr. Salman Naseem Adil as President Elect
Dr. Mehreen Ali Khan as Secretary/Treasurer

# ABOUT PSH

New Executive Committee was elected during 21st Annual Conference of Pakistan Society of Haematology (PSH) held at Karachi from 14th - 16th March 2019. Following are the office bearers of Executive Committee:

Dr. Parvez Ahmed
**President**
Cell: +92 300 8561288
Email: parvez101@yahoo.com

Dr. Salman Naseem Adil
**President Elect**
Cell: +92 300 9249027
Email: salman.adil@aku.edu

Dr. Mehreen Ali Khan
**Secretary/Treasurer**
Cell: +92 333 5164941
Email: mehreen35@hotmail.com

## EXECUTIVE MEMBER COUNSEL:

**Armed Forces**
Qamar Un Nisa Chaudhry
Asad Mahmood Abbasi
Muhammad Sajid Yazdani

**Islamabad**
Nadeem Ikram

**Punjab**
Saima Farhan
Muniza Junaid
Muhammad Irfan Khan

**Sindh**
Syed Muhammad Irfan
Ikram Din Ujjan
Muhammad Nadeem

**Baluchistan**
Hayat Ullah

**Khyberpakhtunkhwa**
Shahtaj Masood

**Azad Kashmir**
Zahida Qasim

**Office Coordinator / IT Manager**
Imran Waheed
Tel: +92 51 9270076 Ext. 228 Mob: +92 322 5181302
Email: psh.org.pk@gmail.com

# ABOUT PSH

| RAWALPINDI/ISLAMABAD | KARACHI | LAHORE |
|---|---|---|
| **Dr Asad Mehmood Abbasi**<br>asadabbasi739@yahoo.com<br>03215506566 | **Dr Bushra Moiz**<br>bushra.moiz@aku.edu<br>0300-2160765 | **Dr Muneeza Junaid**<br>dr.mjunaid10@gmail.com<br>0333-8029026 |

| QUETTA | PESHAWAR | AZAD KASHMIR |
|---|---|---|
| **Dr Hayat Ullah**<br>hayatdotani@yahoo.com<br>03337520574 | **Dr Shahtaj Masood**<br>shahtajmasood@yahoo.com<br>0333-9118335 | **Dr. Zahida Qasim**<br>dr_qasim2011@hotmail.com<br>0333-5875122 |

## SCHEDULE OF MONTHLY MEETING:

| RAWALPINDI / ISLAMABAD | |
|---|---|
| Date: | Last Thursday of the Month |
| Time: | 03:00pm to 05:00pm |
| Coordinator: | Dr Asad Mahmood Abbasi |
| Venue: | AFIP, Rawalpindi |

| KARACHI | |
|---|---|
| Date: | Last Friday of the Month |
| Time: | 08:00am to 09:00am |
| Coordinator: | Dr Bushra Moiz |
| Venue: | AKUH, Karachi |

| LAHORE | |
|---|---|
| Date: | 2nd Tuesday of the Month |
| Time: | 09:00am to 10:00am |
| Coordinator: | Dr Muneeza Junaid |
| Venue: | CIP, Lahore |

| QUETTA | |
|---|---|
| Date: | Last Friday of the Month |
| Time: | 09:00am to 10:00am |
| Coordinator: | Dr Hayat Ullah |
| Venue: | RMC, Quetta |

| PESHAWAR | |
|---|---|
| Date: | 3rd Thursday of the Month |
| Time: | 12:00pm to 01:00pm |
| Coordinator: | Dr Shahtaj Masood |
| Venue: | HMC, Peshawar |

# LT GEN MUHAMMAD SALEEM (1937-2019)

MBBS; DCP (Pb); FRCPath (London); FCPS (Pak); FRCP (Edinburgh)

## CITATION

**By Maj Gen (Retd) Suhaib Ahmed, HI(M)**

On 24 Dec 2019 Army Medical Corps lost a bright and distinguished star named Lt Gen Muhammad Saleem, HI (M) (R). He was a rare breed of outstanding human and professional qualities.

Lt Gen Muhammad Saleem (R) was born at Delhi on 14 Dec 1937 where he received his initial education. In 1947 he migrated to Pakistan along with his parents and the other family members. He did MBBS from Nishtar Medical College in 1959. After doing house job at Nishtar Hospital he joined Army Medical Corps in Apr 1960. He did Grading in Pathology in 1962 and DCP in Pathology in 1966. During the early period of service he remained posted at various field medical units. In 1969 he went to the UK where he worked as registrar Pathology at Dryburn Hospital, Durham and Queen Elizabeth Hospital Birmingham. In 1972 he returned back to Pakistan and established the first department of Haematology at AFIP Rawalpindi. He did MRCPath in Haematology in 1975. There onwards he worked hard to develop the department of Haematology at AFIP. He also served at Saudi Arabia for three years. He worked tirelessly to develop the Haematology Department at AFIP on modern lines. The Department became a reference centre for diagnostic haematology and postgraduate training. He started the first training program for FCPS Haematology in Pakistan. He also started MPhil training in haematology at AFIP.

In the mid-eighties Gen Saleem, under the leadership of Late Lt Gen Syed Azhar Ahmed undertook the construction of Advanced Diagnostic and Research Centre (ADRC) at AFIP. In the late eighties Gen Saleem took keen interest in developing the country's first cytogenetics and molecular genetics services at AFIP. The first clinical service for prenatal diagnosis of thalassaemia and other genetic disorders in Pakistan was started at AFIP in 1994 under the leadership of late Gen Muhammad Saleem. In the early nineties he conceived the idea of establishing a bone marrow transplant centre. He took the initiative to get rotational training of FCPS haematology trainees in general medicine and clinical haematology. He supervised the training of over two

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# CITATION

dozen students in FCPS and MPhil in haematology. He arranged overseas training opportunities for a large number haematologists from the Armed Forces. All of the present day senior haematologists from the Army Medical Corps and most of the senior haematologists from the civilian setups in Rawalpindi and Islamabad are his students.

In 1992 Gen Saleem was promoted to the rank of Maj Gen and was posted to Army Medical College as Professor of Pathology. He came back to AFIP as Commandant in 1994. During his tenure as Commandant, AFIP witnessed continuing progress and improvement. In 1996 he was promoted to the rank of Lt Gen and was appointed Surgeon General and Director General Medical Services. On retirement from Army in Dec 1997 he was appointed as Executive Director National Institute of Health (NIH), Islamabad where he worked hard on the indigenous development of vaccines in Pakistan. In 1997 he also became the founder president of Pakistan Society of Haematology (PSH).

Late Gen Saleem was a great teacher with exceptional communication skills. His passion for teaching and helpful attitude towards students and juniors had earned him a unique honor and respect all over Pakistan.

He always encouraged his students to work on innovative ideas and undertake new research and development projects. He was a gentleman with a very kind heart. He was a very lively personality with a great sense of humor and actively participated in extracurricular activities. He would always be remembered as a dynamic leader and a great mentor. His contributions towards the development of haematology in Pakistan would be remembered in golden words.


بسم اللہ الرحمٰن الرحیم

میں، فاروق احمد خان لغاری

بحیثیت صدر اسلامی جمہوریۂ پاکستان

میجر جنرل محمد سلیم

کو نمایاں عسکری خدمات کے اعتراف میں

ہلالِ امتیاز (عسکری)

کا اعزاز عطا کرتا ہوں۔

مقام: اسلام آباد

تاریخ: ۱۶ ربیع الاوّل ۱۴۱۶ھ

۱۴ اگست ۱۹۹۵ء

**Maj Gen (Retd) Suhaib Ahmed, HI(M)**
CEO, Genetics Resource Centre (GRC), Rawalpindi

# HIGHLIGHTS


The 15th FCPS intensive course was conducted at the The Children's Hospital and Institute of Child Health in collaboration with The College of Physicians and Surgeons of Pakistan and The Shaukat Khanum Memorial Cancer Hospital and Research Centre at 13th-16th November 2019.

Guest of honor was Prof. Abdul Hayee, ex-president of Pakistan Society of Haematology. Formal opening of the course from Maj Gen Parvez Ahmed (Retd), PSH President and welcome addressed by Prof. Masood Sadiq, Dean, The Children's Hospital and Institute of Child Health. It was held under Prof Dr Nisar Ahmed with Dr Saima Farhan being the Chief coordinator and Dr Sarah Rafi being the course coordinator.

Transfusion Medicine module was conducted on 13th November with talks by eminent speakers from all over Pakistan followed by an extensive practice of the wet part and dry part exercises in similarity to those actually held in the FCPS-II exam.

The second day of the course consisted of the module of Coagulation Medicine which followed a similar pattern with lectures, wet and dry part practice.

The third day of the course consisted of elaborative and stimulating talks and lectures on benign and malignant hematology.


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# HIGHLIGHTS


The last day of the course was held at Allama Iqbal Medical College in the department of Hematology in collaboration with the Shaukat Khanum Memorial Cancer Hospital and Research Centre. Elaborative hands on microscopy cases were presented to the candidates followed by a detailed discussion of each case by experts on morphology from all over Pakistan.Overall it was a great learning opportunity and we hope that all the candidates benefitted greatly from it.

# HIGHLIGHTS

HAEMCON 2020

INNOVATION IN HAEMATOLOGY - NEXT PHASE OF INTEGRATED PATIENT CARE
22nd Annual PSH Meeting & International Haematology Congress
February 13th -16th, 2020 | Avari Hotel, Lahore-Pakistan

## PRE-CONFERENCE WORKSHOPS

**Pre-Conference Workshop on "Platelets Disorders and Haemoglobinopathies"** was held at Khawaja Muhammad Safdar Medical College, Sialkot. Dr Muhammad Faisal Bashir and Colonel Rehan Ahmed Lodhi briefed the audience about platelets disorders and haemoglobinopathies.


**Pre-Conference Workshop on "Myeloproliferative Neoplasms"** was held from the platform of PSH in PGMI Hall, Sandeman Provincial Hospital Quetta on 14th December 2019 on the topic of "Myeloproliferative Neoplasms" in which President PSH Maj.General (R) Parvez Ahmed participated as Chief Guest. Symposium was attended by 80 participants. The symposium began with the recitation of The Holy Quran. Porf. Hanif Mengal welcomed the chief guest and the participants. After the welcome address, talks were delivered on Polycythemia vera, Essential Thrombocythemia, Chronic Myeloid Leukemia and Primary Myelofibrosis. Prominent speakers of the symposium were Maj. General (R) Parvez Ahmed (President PSH), Prof. Nadeem Samad Sheikh (Professor of Haematology), Prof, Syed Khalid Shah (Professor of Medicine) and Dr. Hayat Ullah Dotani (Consultant Clinical Haematologist). After the talks, shield distribution ceremony was carried out. At the end of the session, Prof. Chandi Kapoor thanked the Chief Guest and all the participants.

# HIGHLIGHTS

**Pre-Conference Workshop on "Management of Thalassemia"** was held in Khyber Girls Medical College, Peshawar.

Maj. Gen Retd. Prof. Dr. Suhaib Ahmed, HI(M), Former Commandant of AFIP and CEO of Genetics Resource Center, Rawalpindi spoke at length on the approach to the problem of thalassemia in Khyber Pakhtunkhwa as a public health issue from a preventive perspective. He advocated a low cost approach to the problem by introducing a novel 'One Tube Osmotic Fragility Testing' as a qualitative screening test in individuals from families with thalassemia. His team has tested and found the test to have excellent reliability and reproducibility in varied settings. He also introduced a low cost cellulose acetate based hemoglobin electrophoresis equipment. An app called "Thal–IT" can receive and interpret results from the equipment giving the user feedback in real time with a minimal reporting time for patients. Training opportunities in the techniques can be explored individually or by institutions at the Genetics Resource Center at Rawalpindi.

Dr. Nadeem Ikram, Assoc. Professor of Hematology at Rawalpindi Medical College, then, elaborated on the management options in transfusion dependent thalassemia with a focus on optimization of therapy in adolescent thalassemics. Complications of supportive transfusion and iron overload, and their management were discussed to the satisfaction of listening audience.

The last speaker of the day was Maj Gen Prof. Dr. Parvez Ahmed, HI(M) (Retd). He provided the participants with an update on recent trends in bone marrow transplantation in thalassemia patients drawing on his extensive experience in this area at the AFBMTC Rawalpindi and QIH, Islamabad. Dr. Parvez Ahmed emphasized timely consultation with experts in the matter pertaining to patient age and the elements of testing protocols for HLA matching of recipients to various categories of donors.

Prof. Dr. Shahtaj Khan concluded the proceedings of the day by emphasizing the central message of the day, that vital and scarce resources should be allocated by government and non-profits in the future to outreach engagement with families that have high incidence of thalassemia. Investing in counseling services to these families appears to be the only way to put a dent in the rising burden of thalassemia in our region.

# HIGHLIGHTS

**Pre-Conference Workshop on "Haematological Disorders in Pregnancy"** was held in Liaquat University of Medical and Health Sciences (LUMHS) Jamshoro Hospital – at Diagnostic Research Lab Lecture Hall, Hyderabad on 18th December, 2019 in collaboration with Pakistan Society of Hematology (PSH). Moderator for workshop was Dr Mehwish Imam, an Haematology Resident. After registration and sitting of Guests the first session began with recitation from Holy Quran. Prof Dr Ikram Din Ujjan, Dean of Basic medical sciences and Head of Department of Pathology s of LUMHS Jamshoro welcomed the guest speakers and the participants. The Chief Guest of the Symposium was Prof Dr Sohail Almani, who is the Dean of Faculty of Medicine Department and Director of Postgraduate Programs of LUMHS Jamshoro. He addresses the participants and was pleased on having Pre-conference workshop with ultimate success. He further said that such events should be held frequently so that sharing of knowledge stays persistent amongst various institutes. The first lecture presentation was delivered by Dr Munira Burhani, Consultant Haematologist from NIBD, Karachi on Diagnosis and approach to management of Thrombocytopenia in pregnancy. This was Followed by Talk on Hemolytic Diseases of Newborn by Dr Anila Rashid, Consultant Haemtologist from AKUH, Karachi.

The Second session was started By Dr Sidra Asad, Consultant Heamatologist From Patel Hospital, who enlightened the participants with approach to diagnosis and management of Bleeding Disorders. The Last Presentation topic was covered by Sulaiman Zubair who was Product Manager of ASTO life Sciences on Pre Analytical Phase and its Impact on Haematological Testing. The workshop was concluded by vote of Thanks addressed by Dr Kiran Amir , Assistant Professor of Pathology LUMHS Jamshoro. The souvenirs and shields were presented to Chief Guest, Speakers and organizers. It was anticipated by Prof Dr Ikram Din Ujjan at the end that such workshops and symposiums should be organized in future as well.

# HIGHLIGHTS

**Pre-Conference Workshop on "Bone Marrow Morphology - Hands on workshop"** was held at Liaquat University of Medical and Health Sciences (LUMHS), Diagnostic and Research Laboratory at Hyderabad on 19th December 2019. The course was organized by department of pathology of LUMHS, Jamshoro in collaboration with Pakistan Society of Hematology (PSH). The workshop itself was a unique experience for residents as they were exposed to extensive morphology and clinical cases. After sitting of the guest, Professor Dr Farzana Memon who is the Chairperson of the Basic Medical Sciences and Consultant Histhopathologist at LUMHS Jamshoro formally welcomed the guest speakers and participants. The First Session was started by Dr Arsalan Ahmed, Consultant Histopatholgist and Hematopathologist and Section Head of Histopathology of AKUH, Karachi on Flowcytometric diagnosis of B-Cell Lymphoproliferative disorders. After this session ended, all the participants were served with tea and during this time, participants meet the experts of hematology and discussed clinical cases.

The second session started with Bone marrow morphology- Various cases from NIBD and LUMHS Jamshoro were Facilitated and discussed by Dr Syed Jawad, Consultant Hematologist from NIBD, By Dr Kiran Amir and Dr Maryam Khanzada, both being Consultant Haematologists from LUMHS, Jamshoro. At the end, Professor Dr Ikram Din Ujjan - Dean of Basic Medical Sciences and Head of Department of Pathology LUMHS, Jamhoro thanked all the distinguished guests and distributed shields & certificates of appreciation to all speakers, organizers and facilitators

# HIGHLIGHTS

## Seminar-Introduction to AACC Learning Lab (Personalized E-Learning):

The seminar on the AACC Learning Lab held on 26th October, 2019 at Children's Hospital, Lahore was organized by Rahila Institute of Laboratory Medicine as a part of their campaign to promote laboratory teaching across the country. This event was planned in collaboration with Pakistan Society of Hematology (PSH) and The Children's Hospital and Institute of Child Health. The objective of the seminar was to explain how and why Learning Lab can revolutionize the modern lab teaching with its unique way of communicating knowledge. Audience from different levels of experience and a number of institutes turned up and explored the Learning Lab insights. Chief Guest, Dean of faculty Prof. Dr. Masood Sadiq and Head of pathology Dr. Nisar Ahmed expressed their reviews about the learning lab with the latter than expressing his interest in incorporating the teaching tool in PSH curriculum. Keynote speeches are Introduction to AACC Learning Lab by Dr Neelum Mansoor and Collaboration of Pakistan Society of Hematology & Rahila institute of Laboratory Medicine by Dr. Nisar Ahmed.

Event started with Welcome address by Dr. Nazish Saqlain followed by the talk on the agenda of seminar by Dr. Omer Javed from Rahila Institute of Laboratory medicine who explained that how the teams of Learning Lab will be working across the country. An introductory video of the AACC Learning Lab shared in the session followed by a recorded video briefing of Dr. Nader Rifai (Founder of learning lab) highlighted the objectives of learning lab program, course designing and his idea about the future of laboratory learning.

165 participants from 19 different institutions attended the seminar. An attendee survey in the beginning and audience feedback survey at end of program was conducted. Overall this seminar was a great success as audience from all levels were eagerly participated. Dr. Omer concluded the session by satisfying the audience for their queries regarding costing, customized modules and utility of learning lab. He shared his plan of capacity building of lab professionals through hands on skills training and a variety of learning activities throughout the country in collaboration of national bodies. A warm thanks to Dr. Nisar Ahmed and his team who helped in arranging the event and logistics. The management of Rahila institute of laboratory medicine is thankful for Dr. Nisar's cooperation in bringing together experts from all over Lahore to this seminar and encouraging them to participate in this activity.

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# EXTRACORPOREAL PHOTOPHERESIS (ECP)

By Lt Col Ghassan Umair Shamshad, Consultant Haematologist, AFBMTC/NIBMT, Rawalpindi

November 15th 2018 was another landmark day in the history of AFBMTC when the first Extracorporeal Photophoresis (ECP) procedure was performed in any transplant centre of Pakistan. A 20-year-old female who developed chronic Graft-versus-Host Disease following haplo-identical hematopoietic stem cell transplantation (HSCT) for acute myeloid leukemia (AML) was the first patient for this procedure. An off-line ECP machine, Macogenic G2 (Macopharma; Briogene) was installed at AFBMTC, Rawalpindi in November 2018. The off-line ECP procedure has the advantage of being safe for paediatric and <35 kg weight patients in addition to adults. ECP is a very promising approach to treat Graft-versus-host disease (GvHD) following allogeneic HSCT. It is one of the best second-line therapies after failure of steroid treatment, showing a very high therapeutic index and avoiding additional immunosuppression.

**Principle:**

Therapeutic ECP involves the collection of peripheral blood leukocytes by apheresis, extracorporeal exposure of the leukocytes to 8-methoxypsoralen (8-MOP) followed by irradiation with ultraviolet A (UVA) light, and reinfusion of the photoactivated cells. The therapeutic effect of ECP for GvHD appears to be triggered by the ex vivo treated lymphocytes, which undergo apoptosis and modulate a number of in vivo immune responses. These include: increased dendritic cell differentiation; down regulation of autoreactive B cells; alterations in T helper subset populations and lymphocyte homing antigen display; a switch from pro-inflammatory to anti-inflammatory cytokine production; and generation of regulatory T cells.

**Response Rates:**

Overall response rates for steroid-refractory acute GvHD reportedly range from 52–100%; with responses in skin, GI tract and liver ranging from 66–100%, 40– 83%, and 27–71%, respectively. Complete responses and improved survival are often reported among acute GvHD cohorts. Whereas, roughly 30–65% of steroid-dependent patients with chronic GvHD improve with ECP; but most are partial responses. Skin, oral and ocular chronic GVHD manifestations respond in 30–100% of cases while liver, joint, and GI complications improve in 30–80%, 50% and 0–50%, respectively. Duration and frequency of procedures for acute GvHD is once weekly until disease response and then tapered to every-other-week before discontinuation. For chronic GvHD one cycle weekly (or consider twice weekly if treating only mucocutaneous chronic GvHD) until either a response or for 8–12 weeks, followed by a taper to every 2–4 weeks until maximal response.

**Additional Uses:**

In addition to its utility in treating GvHD, ECP is first-line treatment in Cutaneous T-cell lymphoma (Sezary syndrome, Mycosis fungoides) and Bronchiolitis obliterans syndrome. It has also been used in Scleroderma, severe atopic dermatitis of long-standing duration, refractory Crohn's disease and erosive oral lichen planus.

# ROSAI DORFMAN DESTOMBES DISEASE | CASE REPORT

By Dr Maryam Khan, Resident, AFBMTC/NIBMT, Rawalpindi

A 60 years old lady, resident of Chakbeli, Rawat presented with a four months history of bilateral cervical masses, hemoptysis and weight loss. She was a non-smoker with no previous co morbidities. She was a widow and depended on her son for financial support who was a farmer himself and could barely make ends meet.

On physical examination, she had a thin lean body habitus, bilateral upper cervical and preauricular lymphadenopathy (3x3cm). There was no organomegaly .Her blood counts showed anemia (Hb10.4g/dl) and peripheral film showed moderate anisopoikilocytosis, microcytosis and hypochromia. She did not have her baseline LFTs and renal functions assessed at the time of clinic visit. Her cervical lymph node biopsy done from Armed Forces Institute of Pathology, Rawalpindi on 23/07/19 had the following morphological description:

The section reveals a lymph node showing distortion of architecture by dilated sinuses. These inuses show predominantly histiocytes with admixtured lymphocytes, plasma cells and neutrophils. Histiocytes have clear to eosinophilic cytoplasm and showing emperipolesis. Immunohistochemistry revealed S 100 positivity in histiocytes along with CD68 positivity, whereas, CD1a was negative. Based on above findings, an opinion of Rosai Dorfman Destombes Disease was made. Her HRCT chest showed a lytic lesion in right scapula, a soft tissue nodule in upper lobe of left lung and mediastinal lymphadenopathy. MRI Brain showed white matter microvascular angiopathic changes in her brain. On her first clinic visit, she was advised staging bone marrow, viral serology and echocardiography. Her bone marrow aspirate was essentially within normal limits and revealed a few histiocytes and mild hemophagocytosis. A solitary lymphoid follicle consisting of mature lymphocytes accompanied by small collection of histiocytes was seen in trephine biopsy. Immunohistochemistry was not performed owing to financial constraints on behalf of patient. Later on, she was lost to follow up.

**Literature Review:**

Rosai Dorfman Destombes disease is a rare Non-Langerhans cell histiocytosis. The characteristic lesions are positive for S-100 and CD68, but negative for CD1a. The megakaryocytes and histiocytic cells demonstrate varying degrees of emperipolesis. Prevalence is 1:200,000 with an incidence of 100 cases/year in United States. It is more common in males and children and young adults. It is a non-neoplastic disorder that has association with viral infections like EBV, HHV-6, HHv-8, CMV and HIV. It is also associated with mutations of MAP-ERK pathway. Germline mutations of SCL29A3 are frequently seen in familial Rosai Dorfman Destombes Disease (RDD). The disease is also reported in association with Hodgkin Lymphoma, Non-Hodgkin Lymphoma, MDS, Post-allogenic BMT for acute leukemia, cutaneous clear cell sarcoma and concurrent with other histiocytosis. RDD can co-exist with SLE, juvenile Rheumatoid arthritis, autoimmune hemolytic anemia and ALPS. Some extra nodal cases are associated with IgG4 disease. Immunohistochemistry usually shows cytoplasmic and nuclear S100 +ve, fascin +ve; with variable CD68, CD163 and CD14 positivity. CD 1a and langerin are negative. BRAF V600E mutation and Birbeck granules, both found in Langerhans cell histiocytosis, are negative in RDD. It usually has classic nodal presentation with bilateral massive, painless lymphadenopathy. B symptoms may be present. Whereas, extra nodal

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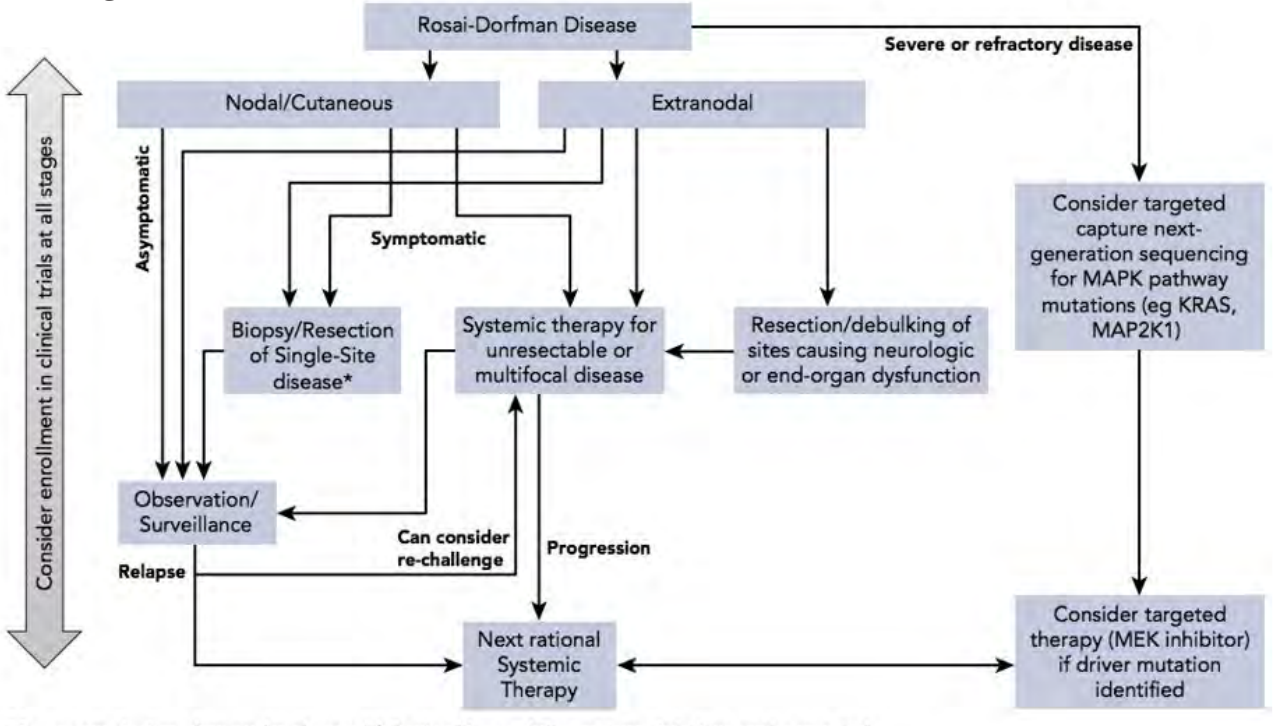
involvement occurs in 43% cases of RDD. Skin is involved in 10% - as painless maculopapular lesions or nodules. There is intracranial, spinal, ophthalmic involvement in <5% cases. Nasal cavity, paranasal sinuses and oral cavity can be involved. Pulmonary involvement is rarely seen. Renal and testicular involvement is also seen usually as renal mass.

**Treatment Options:**

Spontaneous remission is often observed; therefore, the "watch and wait" approach is recommended. It is believed that 70% to 80% of patients have spontaneous improvement of symptoms without treatment. Majority of patients do not require treatment unless the disease is involving vital organs/systems or if the lymph node masses are obstructing airways or spinal cord. The main method of treatment is surgery. For patients requiring systemic treatment, first-line therapeutic option (both in nodal and extranodal RDD) is steroids. However, no standard guidelines regarding the duration and dose are available due to rarity of the disease. Chemotherapy is used in patients with disseminated disease, who do not respond to other therapeutic methods. A number of chemotherapeutic agents have been used which includes vinca alkaloids, alkylating agents, anthracyclines, cladribine, methotrexate, mercaptopurine and azathioprine. Data regarding use of Interferon α, thalidomide, rituximab, imatinib, and retinoids is also available. In steroid resistant patients radiotherapy or newer agents like cobimetinib (MEK inhibitor) appear to be good alternatives. No standard doses of radiotherapy have been established, but doses between 30 and 50 Gy have been employed.

**Treatment Algorithm:**




Treatment Alogrithm - Consensus recommendation for diagnosis and management of Rosai Dorfman Destombes disease. Blood 2018

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# CONGENITAL AMEGAKARYOCYTIC THROMBOCYTOPENIA

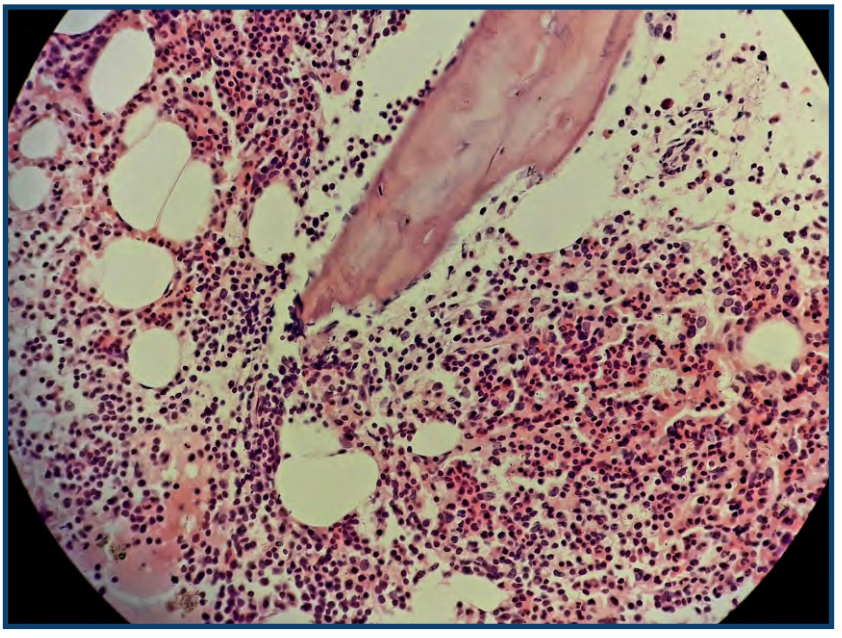
CASE REPORT

By Dr Fayyaz Hussian, Resident, AFBMTC/NIBMT, Rawalpindi

A 3 and a half years old girl was referred to hematology out-patient with a history of fever, recurrent episodes of epistaxis and bleeding from the gums over the past six weeks. She had received platelet transfusions to control bleeding as her platelet count was low and, considering immune mediated destruction as a cause of thrombocytopenia, a trial of glucocorticoids was also given. Her platelet count, however, failed to improve. At the time of referral her fever and bleeding had settled. She was vitally stable, alert and oriented. Bruises and petechiae were noted on lower limbs. No organomegaly or lymphadenopathy was found. Inquiry into past history revealed that the child had experienced rectal bleeding at the age of one year and was found to have low platelet count at that time. She had been provisionally treated as immune thrombocytopenia and received oral glucocorticosteroids for six months. The maximum platelet count she had achieved with steroid therapy was $60 \times 10^9$/l. Till now, there had been no further episodes of bleeding.

Basic labs were ordered including a complete blood count, coagulation profile and routine chemistry. Apart from a low platelet count ($14 \times 10^9$/l) and low Hb (7.6 g/dl), the rest of the investigations were within normal limits. Function studies didn't suggest a platelet function disorder. Given the early age presentation and paucity of response to steroids, a detailed family history was sought which provided an insight into the hereditary nature of the thrombocytopenia as her younger brother also had a history of spontaneous bruising and low platelet count (in range of 30-40 $\times 10^9$/l). Another sibling had died at the age of 02 years due to a bleeding diathesis, the cause of which remained unknown. The remaining 6 siblings (out of total 9) are asymptomatic.

Bone marrow examination of the patient was advised which showed moderately cellular marrow with normal erythropoiesis and myelopoiesis, a relative abundance of histiocytes and foamy macrophages and virtual absence of megakaryocytes in both aspirate and trephine biopsy specimen. Routine cytogenetics revealed a normal female karyotype. Bone marrow examination of the younger affected sibling was done in which megakaryocytes were present but markedly reduced in number. Based on the above findings from clinical and family history and bone marrow examination, a final diagnosis of congenital amegakaryocytic thrombocytopenia was made.



**Discussion:**

Congenital amegakaryocytic thrombocytopenia (CAMT) is a rare autosomal recessive disorder that presents early in life with severe thrombocytopenia. It is a bone marrow failure syndrome which can

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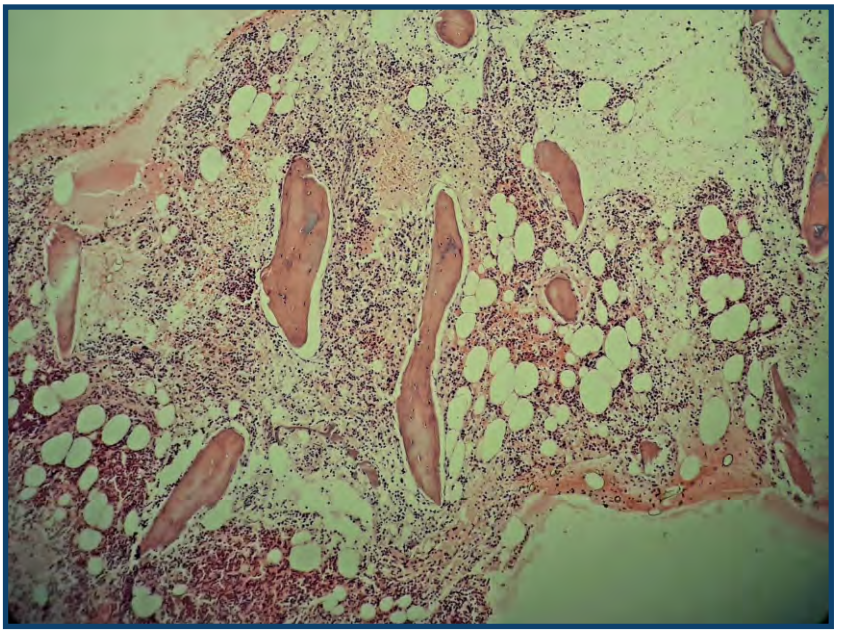
evolve into aplastic anemia and acute leukemia[1]. It routinely presents within the first month of life and is often confused with neonatal alloimmune thrombocytopenia. The other causes of hereditary thrombocytopenia include Wiskot-Aldrich syndrome (WAS) and TAR syndrome (thrombocytopenia with absent radii syndrome). In the absence of genetic studies, the presence of characteristic clinical features (such as skin changes in WAS and skeletal deformities in TAR), variations in platelet size and bone marrow findings are usually sufficient to narrow down a diagnosis. Patient with CAMT can present with muco-cutaneous, gastrointestinal, pulmonary, or intracranial hemorrhage. Thrombocytopenia with megakaryocytopenia is the diagnostic hallmark of the disease. Platelet size is usually normal[2]. It should be noted that megakaryocytes may be normal in number during the first year of life. This does not exclude CAMT as a cause of childhood thrombocytopenia[3]. Mutations in c-Mpl, the gene for thrombopoietin receptor, is present in the majority of patients. Based on clinical progression and genetic findings, a classification scheme was proposed in 2005 which describes three types of CAMT [4,5]:

1. Type 1: Severe thrombocytopenia with early onset bone marrow failure. There is complete loss of functional c-Mpl.
2. Type 2: There is partially functioning c-Mpl gene in these patients. Thrombocytopenia is milder and there may be transient normalization of platelet count during the first year of life. Progression to bone marrow failure occurs relatively late (3-6 years or later).
3. Type 3: In this type of CAMT, there is no defect in c-Mpl gene.

Treatment options are limited. Supportive care with platelet transfusions, fibrinolytic agents in case of minor bleeding, and avoidance of NSAIDs and aspirin are advised. DDAVP may be useful in older children and adults, but should be avoided in infants due to the risk of hyponatremia. ITP-like therapies including corticosteroids and immunoglobulins are ineffective. Newer treatment modalities, such as TPO-mimetics are under investigation. Currently, hematopoietic stem cell transplant is the only curative treatment though it has its attendant mortality and morbidity[6].



**Conclusion**:
Congenital amegakaryocytic thrombocytopenia should be one of the differential diagnoses of childhood thrombocytopenia, especially if it occurs very early in life, there is a family history of

# CASE REPORT

thrombocytopenia or bleeding disorder, or platelet count fails to respond to conventional therapies (e.g Steroids). The absence of physical abnormalities, hereditary nature of the disease and megakaryocytopenia on bone marrow examination favour the diagnosis of CAMT.

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# UPCOMING EVENTS

The 60th Annual Scientific Meeting of the British Society for Haematology,
April 27 - 29, 2020 | ICC Birmingham
https://b-s-h.org.uk/conference-and-events/annual-scientific-meeting/

XXXIII International Symposium on Technical Innovations in Laboratory Hematology
May 21-23, 2020 | Melbourne, Australia - Melbourne Convention and Exhibition Centre
https://www.islh.org/2020/index.php


25th Congress of EHA
June 11 - 14, 2020 | Messe Frankfurt, Germany
https://ehaweb.org/congress/future-congresses/

The 2020 AABB Annual Meeting
October 3-6, 2020 | Baltimore
http://www.aabb.org/annual-meeting/Pages/default.aspx


25th Biennial Annual International Pediatric Conference,
October 23-25, 2020 | Serena Hotel, Quetta
https://www.ppa.org.pk/


18th Biennial International Scientific Conference
October 23-25, 2020 | Karachi
http://sogp.org/conference/

The 62nd American Society of Hematology Annual Meeting and Exposition 2020 (ASH 2020)
December 5-8, 2020 | San Diego, CA, USA
https://www.hematology.org/

# VIEWS & NEWS

The PSH "Haematology Updates" is published on a quarterly basis and is a quick guide to all the happenings in the haematology community.

To improve the updates, your comments and suggestions are welcome.

We further encourage you to send us write ups and photographs of any PSH event in your city/province and they would be featured in our upcoming updates.

For contact, please refer to our corresponding address.

We hope to hear from you on regular basis.

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